REPORT DOCUMENTATION PAGE

Form Approved
OMB No. 0704-0188

PLEASE DO NOT RETURN YOUR FORM TO THE ABOVE ORGANIZATION.

1. REPORT DATE *(DD-MM-YYYY)*	2. REPORT TYPE	3. DATES COVERED *(From - To)*
03/20/2018	Poster	02/15/2018-02/20/2018

4. TITLE AND SUBTITLE
Circumferential Acute Localized Exanthematous Pustulosis

5a. CONTRACT NUMBER

5b. GRANT NUMBER

5c. PROGRAM ELEMENT NUMBER

6. AUTHOR(S)
Capt Pollard, Whitney

5d. PROJECT NUMBER

5e. TASK NUMBER

5f. WORK UNIT NUMBER

7. PERFORMING ORGANIZATION NAME(S) AND ADDRESS(ES)
59th Clinical Research Division
1100 Willford Hall Loop, Bldg 4430
JBSA-Lackland, TX 78236-9908
210-292-7141

8. PERFORMING ORGANIZATION REPORT NUMBER
17506

9. SPONSORING/MONITORING AGENCY NAME(S) AND ADDRESS(ES)
59th Clinical Research Division
1100 Willford Hall Loop, Bldg 4430
JBSA-Lackland, TX 78236-9908
210-292-7141

10. SPONSOR/MONITOR'S ACRONYM(S)

11. SPONSOR/MONITOR'S REPORT NUMBER(S)

12. DISTRIBUTION/AVAILABILITY STATEMENT
Approved for public release. Distribution is unlimited.

13. SUPPLEMENTARY NOTES
American Academy of Dermatology Annual Meeting, San Diego, CA, 15-20 Feb 2018

14. ABSTRACT

15. SUBJECT TERMS

16. SECURITY CLASSIFICATION OF:			17. LIMITATION OF ABSTRACT	18. NUMBER OF PAGES	19a. NAME OF RESPONSIBLE PERSON
a. REPORT	b. ABSTRACT	c. THIS PAGE			Clarice Longoria
					19b. TELEPHONE NUMBER *(Include area code)* 210-292-7141

Approved for public release. Distribution is unlimited.

Circumferential Acute Localized Exanthematous Pustulosis

Whitney L. Pollard, DO, Ngoc Nguyen, BS, Melissa Mauskar, MD
San Antonio Uniformed Services Health Education Consortium, Joint Base San Antonio, TX
University of Texas Southwestern, Dallas, TX

Introduction

Acute localized exanthematous pustulosis (ALEP) is a rare localized variant of acute generalized exanthematous pustulosis (AGEP), first described by Prange et al. in 2005.[1] It is clinically and histologically consistent with AGEP with the exception of its limited distribution on the body. It is characterized by an acute eruption of numerous sterile, coalescing, non-follicular pustules on an erythematous background, most commonly affecting the face, neck and chest.

Contrastingly, AGEP starts in body folds including the axillary, inguinal, and inframammary regions and rapidly spreads to the trunk and limbs within a few hours. Fever, leukocytosis, elevated C-reactive protein, and neutrophilia may accompany the eruption, [2] Systemic symptoms are less common in ALEP. AGEP is drug-induced in 90% of cases, but bacterial and viral infections are also possible culprits.[2, 3] Antibiotics are the most commonly implicated drugs, but others that have been described include anti-hypertensives, antifungals, neuroleptics, analgesics, and NSAIDS.[2] All reports of ALEP have been drug-induced. We present a case of ALEP favored to be secondary to an infectious trigger which has not been previously reported.

Case Presentation

A 16 year-old African American female with a history of atopic triad presented to the emergency department with a pruritic and painful eruption for 2 weeks which started around entirety of hairline and subsequently spread to further involve forehead, neck, and ears. She also endorsed a non-productive cough over the same period. Physical examination revealed sheets of non-follicular pustules on a background of erythema, circumferentially encircling the hairline (Figures 1,2). The helix and post-auricular sulcus were also involved with superficial erosions. She denied a history of new hair products, medications, or known contacts with similar symptomatology. She had been on Claritin, Advair, and Albuterol for a long period of time. A 4 mm punch biopsy was performed which revealed epidermal hyperplasia with neutrophilic spongiosis and a subcorneal pustule in addition to underlying superficial perivascular mixed-cell infiltrate consisting of lymphocytes, neutrophils, and scattered eosinophils. Bacterial culture was negative.

Clinical Presentation

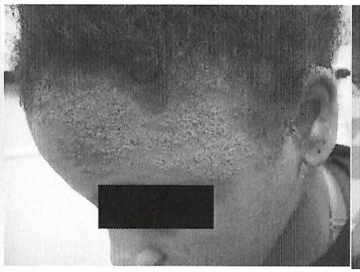

Figure 1

Figure 2

Histology

Figure 3

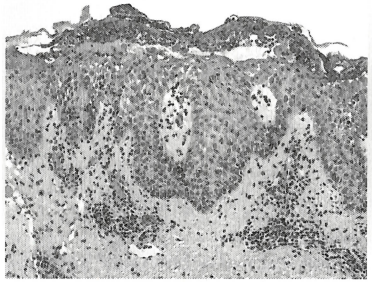

Figure 4

References

1. Prange B, Marini A, Kalke A, Hodzic-Avdagic N, Ruzicka T, Hengge UR. Acute localized exanthematous pustulosis (ALEP). *J Dtsch Dermatol Ges.* 2005;3:210-212.
2. Roujeau JC, Bioulac-Sage P, Bourseau C, Gillaume JC, Bernard P, Lok C, Plantin P, Claudy A, Delavierre C, Vaillant L. Acute generalized exanthematous pustulosis. Analysis of 63 cases. *Arch Dermatol.* 1991;127:1333-8.
3. Chiang SL, Huang YH, Yang CH, Hu S, Hong HS. Clinical manifestations and characteristics of patients with acute generalized exanthematous pustulosis in Asia. *Acta Derm Venereol.* 2008;88:363-365.
4. Britschgi M, Pichler WJ. Acute generalized exanthematous pustulosis, a clue to neutrophil-mediated inflammatory processes orchestrated by T cells. Curr Opin Allergy Clin Immunol 2002;2:325-31.
5. Laurence F, Heidemeyer K, Yawalkar N. Acute generalized exanthematous pustulosis: pathogenesis, genetic background, clinical variants and therapy. *International Journal of Molecular Sciences.* 2016;17:1-9.
6. Langley RGB, Krueger GG, Griffiths CEM. Psoriasis: epidemiology, clinical features, and quality of life. Ann Rheum Dis. 2005;64:18-23.
7. Sneddon IB, WilkinsonDS. Subcorneal pustular dermatosis. *The British Journal of Dermatology.* 1956;68(12):385-394.
8. Tresch S, Cazzo A, Kamarashev J, Harr T, Schid-Grendelmeier P, French LE, Feldmeyer L. T cell-mediated acute localized exanthematous pustulosis caused by finasteride. *J Allergy Clin Immunol.* 2012;129:589-94.
9. Sidoroff A, Halevy S, Bavinck JNB, Vaillant L, Roujeau J. Acute generalized exanthematous pustulosis (AGEP)- A clinical reaction pattern. *J Cutan Pathol.* 2001;28:113-119.

Literature Review & Discussion

The pathogenesis of AGEP/ALEP, though not completely understood, is believed to be T-cell mediated through the production of high levels of cytokines like IL-3 and CXCL8, which recruit and activate neutrophils. Patch tests and in vitro tests have elucidated the important role of drug-specific CD4+ and CD8+ T cells in the development of the condition as these T cells produce significantly more CXCL8 in affected patients.[4] Histology is notable for spongiform subcorneal or intraeptielial pustules, perivascular infiltrates consisting of neutrophils and some eosinophils, focal necrotic keratinocytes, leukocytoclastic vasculitis, and papillary edema.[2, 5]

The differential diagnosis includes:

1. **Pustular psoriasis of von Zumbusch**: This entity is often very difficult to clinically and histologically differentiate from AGEP/ALEP. It is typically triggered by withdrawal of systemic steroids or potent topical steroids and infections, and hospitalization is frequently required.[6]
2. **Sneddon-Wilkinson disease**: The pustules seen in Sneddon-Wilkinson are larger and described as half –and-half vesicles typically sparing the face. The condition most commonly presents in middle-aged women, and rarely occurs in children and adolescents.[7]
3. **Pustular contact dermatitis**: The lack of recent contact with possible allergens or irritants also refutes a diagnosis of pustular contact dermatitis.

There are only a number of case reports describing ALEP, and all of them are drug-related.[8] Besides the chronic use of Claritin, Advair, and Albuterol, this patient did not start any new medications that could explain the eruption. With the history of upper respiratory symptoms accompanying the rash, the likely etiology in this case was a viral infection. Though viral infections have been recognized as a cause of AGEP,[2] to our knowledge, this is the first reported case of non-drug-related ALEP. Treatment includes discontinuing any causative agents and symptomatic relief, including topical steroids for pruritus and antipyretics if it is not a suspected causative drug. The disease is typically self-limited, and the rash can be expected to resolve in less than 15 days, which occurred with topical steroid use in this case.[9]